Research Square

Preprints are preliminary reports that have not undergone peer review.
They should not be considered conclusive, used to inform clinical practice,
or referenced by the media as validated information.

Unri-O Framework for Prognostic Risk Estimation in Children With Hirschsprung Disease: A Pilot Derivation Study

TB Odih Rhomdani Wahid

tubagus7606@lecturer.unri.ac.id

Division of Pediatric Surgery, department of surgery

Annisa' Ulfa

Division of Pediatric Surgery, department of surgery

Ismar -

Division of Pediatric Surgery, department of surgery

Khairuli Amri

Division of Pediatric Surgery, department of surgery

Research Article

Keywords: Hirschsprung Disease, Algorithm, UNRI-O, Risk stratification, Prognostic

Posted Date: November 10th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-11082829/v1

License: This work is licensed under a Creative Commons Attribution 4.0 International License.
Read Full License

Additional Declarations: No competing interests reported.

Abstract

Introduction:

Hirschsprung disease (HSCR) requires individualized diagnostic and surgical management, while postoperative morbidity remains difficult to predict. This study aimed to develop and internally assess the UNRI-O algorithm as a structured framework for diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up.

Methods

This retrospective cohort study included children with HSCR treated at Arifin Achmad General Hospital, Indonesia, from January 2023 to December 2025. Clinical, diagnostic, nutritional, surgical, and postoperative data were analyzed. Binary logistic regression was used to develop the prognostic model, with bootstrap internal assessment, ROC discrimination, and calibration analysis. Variables with $p < 0.25$ or clinical relevance were considered for multivariable modelling.

Results

Of 98 records reviewed, 96 patients comprised the cohort and 95 had complete data for prognostic modelling. Postoperative adverse outcomes occurred in 46 patients. Undernutrition/malnutrition was independently associated with postoperative adverse outcomes, whereas age at surgery was not. The final model showed moderate discrimination (AUC 0.682) and acceptable calibration. An apparent in-sample threshold of 0.411 provided 75.6% sensitivity and 52.0% specificity. Bootstrap assessment indicated substantial variability in coefficient estimates, particularly for nutritional status.

Discussion

Nutritional status was the most informative prognostic factor, supporting nutritional assessment and optimization as part of perioperative HSCR care. UNRI-O integrates prognostic information with diagnostic and surgical considerations into a structured framework.

Conclusions

UNRI-O provides a practical, study-derived framework for risk stratification and perioperative assessment in children with HSCR. External validation in larger multicenter cohorts is required before clinical implementation.

BRIEF POINTS

What is already known on this topic

- Hirschsprung disease (HSCR) requires integrated clinical, radiological, and histopathological assessment for diagnosis.
- Surgical management, including the timing and choice of pull-through technique, is individualized according to disease extent and clinical condition.
- Postoperative complications remain common, while practical tools for individualized prognostic risk estimation in children with HSCR are limited.

What this article adds

- This study developed the UNRI-O algorithm, integrating diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up.
- Nutritional status and age at surgery were incorporated into a study-derived model to estimate the risk of postoperative adverse outcomes, with moderate discrimination.
- UNRI-O provides a structured and practical framework for risk stratification and perioperative assessment in children with HSCR, while further external validation is required.

INTRODUCTION

Hirschsprung disease (HSCR) is a congenital disorder of the enteric nervous system characterized by the absence of ganglion cells in the distal intestine, resulting in functional intestinal obstruction. Although definitive treatment is surgical, postoperative complications and bowel dysfunction remain important clinical challenges, with outcomes varying according to disease extent, clinical condition, and other patient-related factors. Current clinical guidelines provide recommendations for diagnosis, surgical management, and long-term follow-up, but individualized prognostic risk estimation remains limited.[1]

Recent studies have begun to explore prediction models for specific postoperative outcomes, particularly Hirschsprung-associated enterocolitis, but a broader framework integrating diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up is still needed. Therefore, this study aimed to develop and internally assess the UNRI-O algorithm as a structured, study-derived framework for risk stratification of postoperative adverse outcomes in children with HSCR.[2]

METHODS

Study Design, Participants, and Selection Bias

This retrospective cohort study was conducted at Arifin Achmad General Hospital, Pekanbaru, Riau, Indonesia, using medical records of children with HSCR from January 2023 to December 2025. Following ethical approval and authorization to access medical records, no formal a priori sample size calculation was performed. The study sample was a total sampling with all available pediatric HSCR

records during the study period were reviewed for eligibility and completeness. Patients with confirmed HSCR who underwent surgical treatment and had sufficient clinical, diagnostic, surgical, and postoperative data were included. Records with incomplete essential information or loss to follow-up were excluded. Of 98 records reviewed, 96 patients were included in the primary cohort. One patient was excluded because essential supporting-investigation and operative data were unavailable, while another was excluded because of loss to follow-up before surgery and therefore had no operative or postoperative outcome data. To minimize information bias, all eligible medical records were reviewed systematically using predefined study variables and outcome definitions.

Variables and Outcomes

Demographic, clinical, diagnostic, nutritional, surgical, and postoperative data were extracted from medical records. Age at surgery was categorized as neonate (< 28 days), infant (29–365 days), or child (> 365 days). Clinical presentation was assessed according to the number of characteristic HSCR features. Diagnostic assessment included abdominal radiography, contrast enema/colon-in-loop examination, and rectal biopsy, while disease extent was classified as ultrashort, short, long-segment disease, total colon aganglionic (TCA). For prognostic modelling, nutritional status was categorized as good nutrition or undernutrition/malnutrition. Serum albumin was classified as normal, mild, moderate, or severe hypoalbuminemia, or not tested. The primary outcome was postoperative adverse outcome, defined as at least one postoperative complication, including obstructive symptoms, Hirschsprung-associated enterocolitis (HAEC), soiling, or other complications. Mortality was recorded separately.

Statistical Analysis

Categorical variables were summarized as frequencies and percentages. Associations between potential predictors and postoperative adverse outcomes were assessed using Pearson's chi-square test when expected cell counts were adequate and Fisher's exact or Fisher–Freeman–Halton exact tests for sparse data. Variables with $p < 0.25$ in bivariate analysis, together with clinically relevant variables specified a priori, were considered for multivariable modelling. Binary logistic regression was used to develop the prognostic model, with odds ratios (ORs) and 95% confidence intervals (CIs). Model fit was assessed using the omnibus test, Nagelkerke R^2, and Hosmer–Lemeshow goodness-of-fit test. Models with convergence problems due to sparse categories were not retained. The final model included age at surgery and categorized nutritional status. The predicted probability of an adverse postoperative outcome was calculated using the logistic regression equation, where x is the linear predictor, with children (> 365 days) and well-nourished patients serving as the reference categories :

logit(p) = -0.818 + 0.133(Neonate) + 0.765(Infant) + 1.685(Undernutrition/Malnutrition)

$$p = \frac{1}{1 + e^{-x}}$$

Internal validation was assessed using 1,000 bootstrap resamples with bias-corrected and accelerated (BCa) 95% CIs. Discrimination was evaluated using the area under the receiver operating characteristic curve (AUC), and calibration was assessed using the Hosmer–Lemeshow test and comparison of observed and predicted probabilities. An apparent probability threshold was derived using the Youden index and considered exploratory because it was derived from the same cohort used for model development. Missing data categorized nutritional-status and was therefore excluded from complete-case multivariable and ROC analyses, resulting in 95 patients for these analyses. Statistical tests were two-sided, with $p < 0.05$ considered statistically significant.

Development of the UNRI-O Algorithm

The UNRI-O algorithm was developed from the findings of this study as a research-derived risk-estimation framework for children with Hirschsprung disease. It integrates four sequential components: diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up. The prognostic component incorporates age at surgery and categorized nutritional status from the multivariable model to estimate the individual probability of postoperative adverse outcomes. Thus, UNRI-O represents a study-derived risk-estimation framework intended to support clinical assessment and risk stratification estimation.

Ethical Considerations

The study was approved by the Ethical Review Board for Medicine & Health Research, Faculty of Medicine, Universitas Riau. Ethical approval was granted under approval number 081/UN19.5.1.1.8/UEPKK/2026. Authorization to access medical records was obtained following ethical approval. Informed consent was not applicable because this study used secondary data obtained from existing medical records and involved no direct interaction with patients or their parents/guardians. Patient identifiers were removed and replaced with study codes.

RESULTS

Study Population

a total of 98 medical recods of children with HSCR were initially reviewed. Two patients were excluded: one because of incomplete essential diagnostic and operative data and one because of loss to follow-up before surgery, 96 patients were included in the final study cohort. Complete data for the primary prognostic model were available for 95 patinets, as one patient had missing data for categorized nutritional status and was therefore excluded from the complete-case multivariable analysis. The remaining 96 patients were retained in the overall cohort and decriptive analysis.

Baseline and clinical characteristics

The cohort comprised 64 males (66.7%) and 32 females (33.3%). At definitive surgery, most patients were infants (49, 51.0%), followed by children older than 365 days (35, 36.5%) and neonates (12, 12.5%). Short-segment disease predominated (62, 64.6%), and staged surgery was performed in 61 patients (63.5%). Duhamel pull-through was the most commonly used technique (60, 62.5%), followed by TEPT (32, 33.3%). Most patients had good nutritional status (77, 80.2%), while 18 of 95 patients with complete nutritional data (18.9%) were classified as undernourished/malnourished for prognostic analysis.

Table 1
Baseline characteristics of Children with Hirschsprung Disease Included in the Study

Characteristics	N (%)
Age at admission	
Neonate (≤ 28 days)	28 (29.2)
Infant (29–265 days)	41 (42.7)
Child (> 365 days)	27 (28.1)
Age at Surgery	
Neonate (≤ 28 days)	12 (12.5)
Infant (29–265 days)	49 (51.0)
Child (> 365 days)	35 (36.5)
Sex	
Male	64 (66.7)
Female	32 (33.3)
Clinical Triad Components	
Two Components	41 (42.7)
Three Components	55 (57.3)
Supporting Investigations	
1 Investigations	8 (8.3)
2 Investigations	60 (62.5)
3 Investigations	28 (29.2)
X-ray	
Performed and supportive of HSCR	57 (59.4)
Not Performed	39 (40.6)
Contrast Enema (Colon in Loop)	
Performed and supportive of HSCR	94 (97.9)
Not Performed	2 (2.1)
Rectal Biopsy	
Performed and supportive of HSCR	61 (63.5)

Characteristics	N (%)
Age at admission	
Not Performed	35 (36.5)
Aganglionic Segments	
Ultrashort Segment	27 (28.1)
Short Segment (Rectosigmoid)	62 (64.6)
Long Segment	7 (7.3)
Total Colon Aganglionic (TCA)	0 (0.0)
Surgical Approach	
Single-stage primary pullthrough	32 (33.3)
Two-stage	61 (63.5
No data	3 (3.1)
Definitive Pullthrough Technique	
Transanal Endorectal Pullthrough (TEPT)	32 (33.3)
Duhamel Procedure	60 (62.5)
Soave/Soave-Boley Procedure	3 (3.1)
Swenson Procedure	0 (0.0)
No data	1 (1.0)
Serum Albumin Status	
Normal (≥ 3.5 g/dL)	33 (34.4)
Mild Hypoalbuminemia (3.0 - 3.4 g/dL)	14 (14.6)
Moderate Hypoalbuminemia (2.5–2.9 g/dL)	5 (5.2)
Severe Hypoalbuminemia (< 2.5 g/dL)	2 (2.1)
Laboratory test unrecorded	42 (43.8)
Nutritional Status (N = 95 complete)	
Well-nouroshed	77 (81.1) (80.2% total)
Undernourished/Malnourished	18 (18.9) (18.8% total)

Postoperative Outcomes

Postoperative adverse outcomes occurred in 46 of 96 patients (47.9%), comprising Hirschsprung-associated enterocolitis in 20 (20.8%), postoperative obstructive symptoms/constipation in 16 (16.7%), other complications in 8 (8.3%), and soiling in 2 (2.1%). Overall postoperative mortality was 3.1% (n = 3). Mortality was analyzed separately from the composite adverse-outcome endpoint. The three deaths were included within the adverse-outcome group and therefore were not counted as additional adverse events.

Table 2
Postoperative Outcomes characteristics of Children with Hirschsprung Disease Included in the Study

Characteristics	**N (%)**
Adverse Outcome	
Obstructive symptoms pasca pullthrough	16 (16.7)
HAEC	20 (20.8)
Soiling	2 (2.1)
Others	8 (8.3)
No adverse outcome	50 (52.1)
Mortality	
Death	3 (3.1)
Postoperative adverse outcome overall	46 (47.9)

Bivariate Analysis

Bivariate analyses were performed to assess the association between demographic, clinical, diagnostic, disease-related, surgical, nutritional, and laboratory characteristics and postoperative adverse outcomes. categorized nutritional status was the only variable significantly associated with postoperative adverse outcomes, occurring in 31 of 77 (40.3%) children with good nutrition versus 14 of 18 (77.8%) with undernutrition/malnutrition (χ^2=8.237, p = 0.004). No significant associations were observed for the remaining variables, including age at surgery (p = 0.264), segment length (p = 0.284), and pull-through technique (p = 0.803).

Table 3
Clinical and Surgical Characteristics according to Postoperative Adverse Outcome

Variable	No Adverse Outcome (N = 50)	Adverse Outcome (N = 46)	p-value	Statistical Test
Age at admission			0.570	Pearson Chi-square (χ^2=1.124)
Neonate (≤ 28 days)	15 (53.6%)	13 (46.4%)		
Infant (29–265 days)	19 (46.3%)	22 (53.7%)		
Child (> 365 days)	16 (59.3%)	11 (40.7%)		
Age at Surgery			0.264	Pearson Chi-square (χ^2=2.662)
Neonate (≤ 28 days)	6 (50.0%)	6 (50.0%)		
Infant (29–265 days)	22 (44.9%)	27 (55.1%)		
Child (> 365 days)	22 (62.9%)	13 (37.1%)		
Sex			0.666	Fisher's Exact Test
Male	32 (50.0%)	32 (50.0%)		
Female	18 (56.3%)	14 (43.8%)		
Clinical Triad Components			0.152	Fisher's Exact Test
Two Components	25 (61.0%)	16 (39.0%)		
Three Components	25 (45.5%)	30 (54.5%)		
Supporting Investigations			0.660	Fisher-Freeman-Halton Exact
1 Investigations	3 (37.5%)	5 (62.5%)		
2 Investigations	33 (55.0%)	27 (45.0%)		
3 Investigations	14 (50.0%)	14 (50.0%)		
X-ray			0.536	Fisher's Exact Test
Performed and supportive of HSCR	28 (49.1%)	29 (50.9%)		
Not Performed	22 (56.4%)	17 (43.6%)		
Contrast Enema (Colon in Loop)			1.000	Fisher's Exact Test
Performed and supportive of HSCR	49 (52.1%)	45 (47.9%)		
Not Performed	1 (50.0%)	1 (50.0%)		

Variable	No Adverse Outcome (N = 50)	Adverse Outcome (N = 46)	p-value	Statistical Test
Rectal Biopsy			0.399	Fisher's Exact Test
Performed and supportive of HSCR	34 (55.7%)	27 (44.3%)		
Not Performed	16 (45.7%)	19 (54.3%)		
Aganglionic Segments			0.286	Fisher-Freeman-Halton Exact
Ultrashort Segment	11 (40.7%)	16 (59.3%)		
Short Segment (Rectosigmoid)	36 (58.1%)	26 (41.9%)		
Long Segment	3 (42.9%)	4 (57.1%)		
Total Colon Aganglionic (TCA)	0 (0.0%)	0 (0.0%)		
Surgical Approach			0.580	Fisher-Freeman-Halton Exact
Single-stage primary pullthrough	15 (46.9%)	17 (53.1%)		
Two-stage	34 (55.7%)	27 (44.3%)		
No data	1 (33.3%)	2 (66.7%)		
Definitive Pullthrough Technique			0.803	Fisher-Freeman-Halton Exact
Transanal Endorectal Pullthrough (TEPT)	17 (53.1%)	15 (46.9%)		
Duhamel Procedure	32 (53.3%)	28 (46.7%)		
Soave/Soave-Boley Procedure	1 (33.3%)	2 (66.7%)		
Swenson Procedure	0 (0.0%)	0 (0.0%)		
No data	0 (0.0%)	1 (100.0%)		
Serum Albumin Status			0.079	Fisher-Freeman-Halton Exact
Normal (≥ 3.5 g/dL)	18 (54.5%)	15 (45.5%)		
Mild Hypoalbuminemia (3.0 - 3.4 g/dL)	8 (57.1%)	6 (42.9%)		
Moderate Hypoalbuminemia (2.5–2.9 g/dL)	0 (0.0%)	5 (100.0%)		
Severe Hypoalbuminemia (< 2.5 g/dL)	0 (0.0%)	2 (100.0%)		

Variable	No Adverse Outcome (N = 50)	Adverse Outcome (N = 46)	p-value	Statistical Test
Laboratory test unrecorded	24 (57.1%)	18 (42.9%)		
Nutritional Status (N = 95 complete)			0.004	Score Test/Chi-Square (χ^2=8.237)
Well-nouroshed	46 (59.7%)	31 (40.3%)		
Undernourished/Malnourished	4 (22.2%)	14 (77.8%)		

Prognostic Model Development

Multivariable logistic regression was used to develop the prognostic model for postoperative adverse outcomes. Model 3, incorporating age at surgery and categorized nutritional status, was selected as the primary model because it was parsimonious and converged successfully (n = 95). Undernutrition/malnutrition was independently associated with higher odds of postoperative adverse outcomes compared with good nutritional status (OR 5.394, 95% CI 1.537–18.937; p = 0.009), whereas age at surgery was not independently associated with the outcome. The overall model was statistically significant (χ^2=11.427, df = 3, p = 0.010), with a Nagelkerke R^2 of 0.151 and good model fit (Hosmer–Lemeshow p = 0.921). Models incorporating detailed albumin categories or pull-through technique were not retained because of convergence problems related to sparse categories.

Table 4
Multivariable Logistic Regression analysis of Factors associated with Postoperative Adverse Outcome

Variable	β	SE	Wald	p-value	OR	95% CI
Age at surgery						
Neonate vs child	0.133	0.750	0.031	0.860	1.142	0.263–4.964
Infant vs child	0.765	0.477	2.571	0.109	2.150	0.844–5.480
Nutritional status						
Undernutrition/malnutrition vs well-nourished	1.685	0.641	6.919	0.009	5.394	1.537–18.937

Model statistics

N = 95; Omnibus χ^2 = 11.427, df = 3, p = 0.010; Cox & Snell R^2 = 0.113; Nagelkerke R^2 = 0.151; Hosmer–Lemeshow χ^2 = 0.490, df = 3, p = 0.921.

Internal Validation and Model Discrimination

Internal validation of Model 3 was performed using 1,000 bootstrap resamples with bias-corrected and accelerated (BCa) 95% confidence intervals. The original regression coefficient for undernutrition/malnutrition was 1.685, with a bootstrap standard error of 3.000 and a BCa 95% confidence interval ranging from − 0.097 to 27.193. The corresponding bootstrap estimates for the neonatal and infant age categories were also associated with relatively wide confidence intervals. The bootstrap results indicated substantial variability in the coefficient estimates, particularly for nutritional status. Accordingly, the model estimates were interpreted with caution given the limited sample size and sparse distribution of some categories.

Table 5
Boostrap Internal Validation of the Final Prognostic Model

Variable	Original β	Bootstrap SE	Bootstrap p-value	BCa 95% CI
Neonate vs child	0.133	1.983	0.851	−1.613 to 1.712
Infant vs child	0.765	0.519	0.106	−0.276 to 2.111
Undernutrition/malnutrition vs well-nourished	1.685	3.000	0.003	−0.097 to 27.193
Constant	−0.818	0.409	0.029	−1.638 to −0.246

The discriminative performance of Model 3 was assessed using receiver operating characteristic (ROC) analysis based on the model-derived predicted probabilities. The model yielded an area under the ROC curve (AUC) of 0.682 (95% CI 0.574–0.790; SE 0.055; p = 0.001), indicating moderate discrimination between patients with and without postoperative adverse outcomes.

Table 6
Discrimination Performance of the UNRI-O Prognostic Model

Metric	Estimate	SE	95% CI	p-value
AUC	0.682	0.055	0.574–0.790	0.001

An apparent in-sample threshold of 0.411, derived using the Youden index, yielded a sensitivity of 75.6% and a specificity of 52.0%, with a Youden index of 0.236. This threshold was derived from the same dataset used for model development and therefore represents an apparent threshold rather than a validated clinical cutoff.

Model Calibration

Model calibration was assessed using the Hosmer–Lemeshow test and comparison of observed versus predicted probabilities across six groups of predicted risk. The Hosmer–Lemeshow test did not indicate evidence of poor model fit (χ^2=0.490, df = 3, p = 0.921). Across the six predicted-probability groups, observed event rates were generally close to the corresponding mean predicted probabilities, although some groups contained relatively few patients. The predicted probabilities ranged from approximately 30.6% to 83.7% across the combinations of age at surgery and nutritional status. The lowest estimated probability was observed among older children with good nutritional status, whereas the highest was observed among infants with undernutrition/malnutrition.

Table 7
Model Calibration of the UNRI-O Prognostic Model

Predicted probability	**N**	**Observed adverse outcome N (%)**	**Mean predicted probability**
0.30612	30	9 (30.0)	0.3061
0.33500	7	2 (28.6)	0.3350
0.48679	40	20 (50.0)	0.4868
0.70412	4	3 (75.0)	0.7041
0.73100	5	4 (80.0)	0.7310
0.83651	9	7 (77.8)	0.8365
Overall	**95**	**45 (47.4)**	**0.4737**

Hosmer–Lemeshow test: χ^2 = 0.490, df = 3, p = 0.921.

UNRI-O Risk Matrix

Based on the final logistic regression model, a clinically simplified risk matrix was generated using age at surgery and categorized nutritional status, presented in Table 8.

Table 8
Estimated Probability of Postoperative Adverse Outcome according to Age at Surgery and Nutritional Status

Age at surgery	**Good nutrition**	**Undernutrition/malnutrition**
Neonate (≤ 28 days)	33.5%	73.1%
Infant (29–365 days)	48.7%	83.7%
Child (> 365 days)	30.6%	70.4%
Note: Predicted probabilities were derived from the final logistic regression model in the derivation cohort and have not undergone external validation.		

The matrix represents model-derived predicted probabilities of postoperative adverse outcomes for each combination of age at surgery and nutritional status. These estimates were generated from the derivation dataset and have not undergone external validation. The resulting UNRI-O prognostic framework incorporated age at surgery and nutritional status to provide individualized estimates of postoperative adverse-outcome probability. Model performance in the derivation cohort was characterized by moderate discrimination and acceptable calibration, while the observed variability in bootstrap estimates highlighted the uncertainty associated with model coefficients in this relatively small cohort.

Development of the UNRI-O Algorithm

Based on the diagnostic, surgical, and prognostic findings of this cohort, we developed the UNRI-O algorithm as a structured framework for children with Hirschsprung disease. The algorithm comprises four sequential phases: diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up. The diagnostic phase integrates clinical assessment, supporting investigations, histopathological confirmation, and classification of aganglionosis extent. Surgical planning considers disease extent and clinical condition to guide a one-stage or staged approach, with the choice of pull-through technique based on anatomical and clinical considerations rather than the prognostic model. The prognostic phase incorporates age at surgery and categorized nutritional status to estimate the individual probability of postoperative adverse outcomes, with the resulting probabilities presented as a risk matrix. The final phase consists of postoperative surveillance for complications and mortality. The complete UNRI-O framework is presented in Fig. 3.

DISCUSSION

This study developed the UNRI-O algorithm as a structured framework integrating diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up. In this cohort, most patients were infants at definitive surgery, had short-segment disease, underwent staged surgery, and had good nutritional status. Despite these characteristics, postoperative morbidity remained an important clinical concern. Nutritional status emerged as the main prognostic factor, with children who were undernourished or malnourished experiencing a greater risk of postoperative adverse outcomes. This finding is consistent with previous studies showing that poor nutritional status is associated with postoperative morbidity in children with HSCR, particularly Hirschsprung-associated enterocolitis.

Nutritional status may therefore serve not only as a marker of baseline clinical condition but also as a potentially modifiable factor that could be considered during preoperative assessment and postoperative risk stratification. In the final prognostic model, undernutrition/malnutrition was the only independent predictor of postoperative adverse outcomes, while age at surgery was retained as a clinically relevant covariate. The model demonstrated moderate discrimination (AUC 0.682) and acceptable calibration in the derivation cohort. Our finding that undernutrition/malnutrition was associated with approximately five-fold higher odds of postoperative adverse outcomes is consistent with previous evidence linking nutritional status to postoperative morbidity in HSCR. Nutrition

assessment for further adequate nutrition support is one of the crucial aspects in the overall management of HSCR. Illness-related malnutrition is prevalent in up to 40% of surgical patients, undernutrition was identified as an independent factor predicting poor postoperative outcomes in pediatric patients. In patients with HSCR, studies identifying predictors of postoperative Hirschsprung-associated enterocolitis (HAEC) reported that malnutrition was associated with the presence of postoperative recurrent HAEC and a high risk of mortality.[3,4]

In our cohort, most patients underwent definitive surgery during infancy, and the Duhamel procedure was the most commonly used pull-through technique. Age at surgery was not independently associated with postoperative adverse outcomes in our cohort, although it was retained in the final model because of its clinical relevance. This finding suggests that postoperative morbidity may depend more on the patient's underlying clinical condition and nutritional status than on age at definitive surgery alone. These adverse events are often multifactorial, influenced by both surgical technique and timing of intervention or age at surgery.[5] There is no consensus on the optimal age for performing definitive surgery. Some studies suggest that earlier surgery may promote better functional outcomes and reduce the incidence of complications by taking advantage of faster tissue healing and earlier bowel adaptation. Others argue that delayed surgery may be safer or technically easier in older infants due to more mature anatomical structures and a larger body size. As a result, the timing of definitive pull-through surgery remains a matter of ongoing debate.[6]

Similarly, pull-through technique was not significantly associated with adverse outcomes in bivariate analysis and was not retained in the final model because sparse representation of some techniques resulted in convergence problems. This finding should not be interpreted as evidence that different techniques are equivalent. Surgical treatments aim to remove the aganglionic segment and restore bowel continuity. Variations of definitive pull-through remain the primary surgical approach for HSCR. Although historically commonly performed, the Duhamel procedure is now used less frequently in contemporary pediatric practice. Current evidence and guidelines do not favor one technique and recommend that surgeons perform the type of pull-through with which they have the most experience. Short-term postoperative adverse events are relatively common, including Hirschsprung-associated enterocolitis (HAEC), anastomotic leakage, anastomotic strictures and surgical site infections (SSI).[7]

Risk estimation frameworkling in HSCR is an emerging approach for identifying patients at increased risk of postoperative morbidity. Recent studies have developed prediction models targeting specific outcomes, particularly postoperative HAEC, using routinely available clinical and nutritional variables.[2,8] In this context, the UNRI-O algorithm provides a broader framework by integrating diagnostic assessment, surgical planning, prognostic risk estimation, and postoperative follow-up. Its prognostic component translates age at surgery and nutritional status into an individualized estimate of postoperative adverse outcome risk, providing a structured approach to patient risk stratification.[9,10]

Importantly, UNRI-O should currently be regarded as a study-derived risk-estimation framework rather than a validated clinical prediction rule. This study has several limitations, its retrospective, single-center

design and relatively limited sample size may restrict the generalizability and stability of the findings. Internal validation using bootstrap resampling provides an assessment of model stability within the development population, but external validation in independent and preferably multicenter cohorts is required before the predicted risks can be generalized or used to guide clinical decisions. Therefore, further prospective, multicenter studies are needed to confirm its predictive performance and clinical applicability in broader HSCR populations.

CONCLUSIONS

The UNRI-O algorithm is a pilot derivation study that offers a structured framework for risk stratification in children with Hirschsprung disease, with nutritional status emerging as an important prognostic factor. External validation is needed before clinical implementation.

Declarations

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

FUNDING

DISCLOSURE

This study was partially supported by a research grant from Lembaga Penelitian dan Pengabdian Kepada Masyarakat (LPPM), Universitas Riau. The remaining costs were covered by the authors.

Author Contribution

T.O.R: conceptualization, methodoly, writing-original draft preparation, literature review. AU: conceptualization, methodoly, writing-original draft preparation, data collection and results interpretation. I: conceptualization, methodoly, writing-original draft preparation, literature review. KA: methodoly, writing-original draft preparation. All authors have read and agreed to the published version of the manuscript.

Acknowledgement

The authors would like to thank the Department of Pediatric Surgery, Arifin Achmad General Hospital, Pekanbaru, Riau, Indonesia, for their support and assistance throughout this study. We also thank the medical record staff for their assistance in data retrieval and documentation.

Data Availability

The data supporting the findings of this study are available from the corresponding author upon reasonable request, subject to institutional and ethical restrictions. Individual-level data are not publicly available to protect patient confidentiality.

References

1. Kyrklund K, Sloots CEJ, de Blaauw I, Bjørnland K, Rolle U, Cavalieri D et al (2020) ERNICA guidelines for the management of rectosigmoid Hirschsprung's disease. Orphanet J Rare Dis 15:164. https://doi.org/10.1186/s13023-020-01362-3
2. Rossi D, Irvine W, Granström AL, Mutanen A, Bakx R, Bjørnland K et al (2026) Updated ERNICA guidelines for the management of rectosigmoid Hirschsprung's disease 2025. J Pediatr Gastroenterol Nutr. https://doi.org/10.1002/jpn3.70442
3. Zhang HY, Chen K, Zhang Y, Gao T, Huang Y, Ge L et al Preoperative nutritional support in children with Hirschsprung disease: a prospective multicenter open-label randomized controlled trial. Nat Commun 2025;16. https://doi.org/10.1038/s41467-025-66541-x
4. Wang D, Zhu T, Zhu L, Ji C, Zhou B, Zhang G et al (2023) Screening of undernutrition in children with Hirschsprung disease using preoperative anthropometric parameters: A multicenter cross-sectional study. J Parenter Enter Nutr 47:151–158. https://doi.org/10.1002/jpen.2440
5. Poerwosusanta EE, Hariastawa IA, Setiawan A, Sulistiawati S, Ary Wibowo A (2021) Correlation Between The Initial-Surgery Age With The Hirschsprung's Disease Patient Stooling Patterns In Ulin Hospital Banjarmasin, South Kalimantan, Indonesia In 2012–2015. Majalah Biomorfologi 31:66. https://doi.org/10.20473/mbiom.v31i2.2021.66-69
6. Sowulewski O, Leszkowicz J, Sakowska M, Spychalski P, Szlagatys-Sidorkiewicz A (2026) Definitive surgery for Hirschsprung disease between 3 and 12 months achieves best outcomes: A systematic review with meta-analysis. J Pediatr Surg 61. https://doi.org/10.1016/j.jpedsurg.2026.163039
7. Mao Yzhong, tao Tang S, Li S (2018) Duhamel operation vs. transanal endorectal pull-through procedure for Hirschsprung disease: A systematic review and meta-analysis. J Pediatr Surg 53:1710–1715. https://doi.org/10.1016/j.jpedsurg.2017.10.047
8. Jiang Z, Hou J, Feng W, Die X, Liu Q, Guo Z et al (2026) Development and validation of nomogram model for postoperative enterocolitis using LASSO-logistic regression: single-center retrospective study of 424 Hirschsprung disease patients. Eur J Med Res 31:1259. https://doi.org/10.1186/s40001-026-05008-1
9. Zheng W, Chen W, Chen F, Wang Y, Zhu L, Liu M Development of a nomogram for predicting postoperative Hirschsprung-associated enterocolitis. Front Pediatr 2025;13. https://doi.org/10.3389/fped.2025.1640331
10. Die X, Feng W, Song A, Liu W, Wang Y, Guo Z et al (2024) Risk factors and simple scoring system for predicting postoperative nutritional status of Hirschsprung's disease. Front Nutr 11.

https://doi.org/10.3389/fnut.2024.1441104

Figures

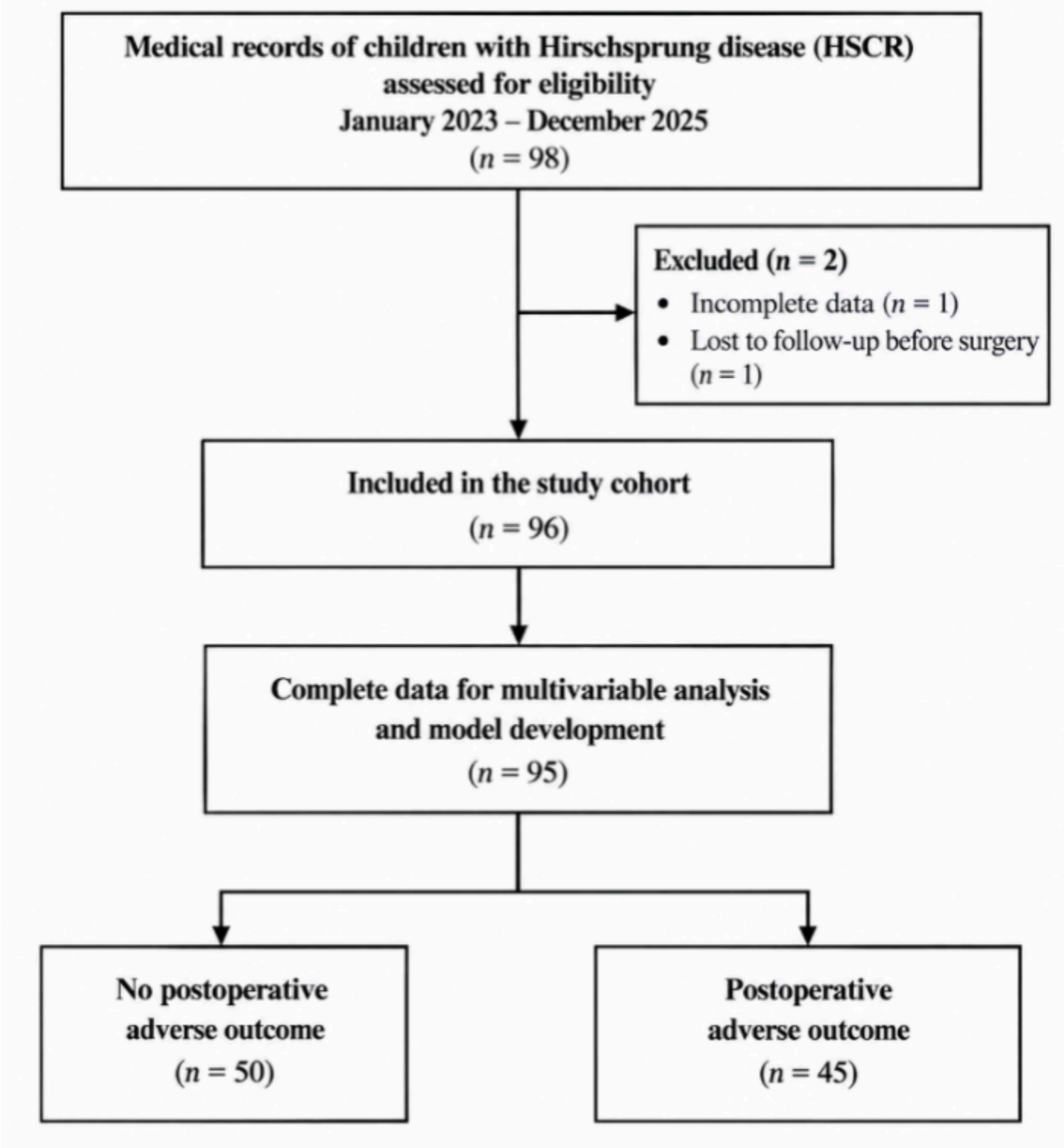


Figure 1

Study Flow Diagram

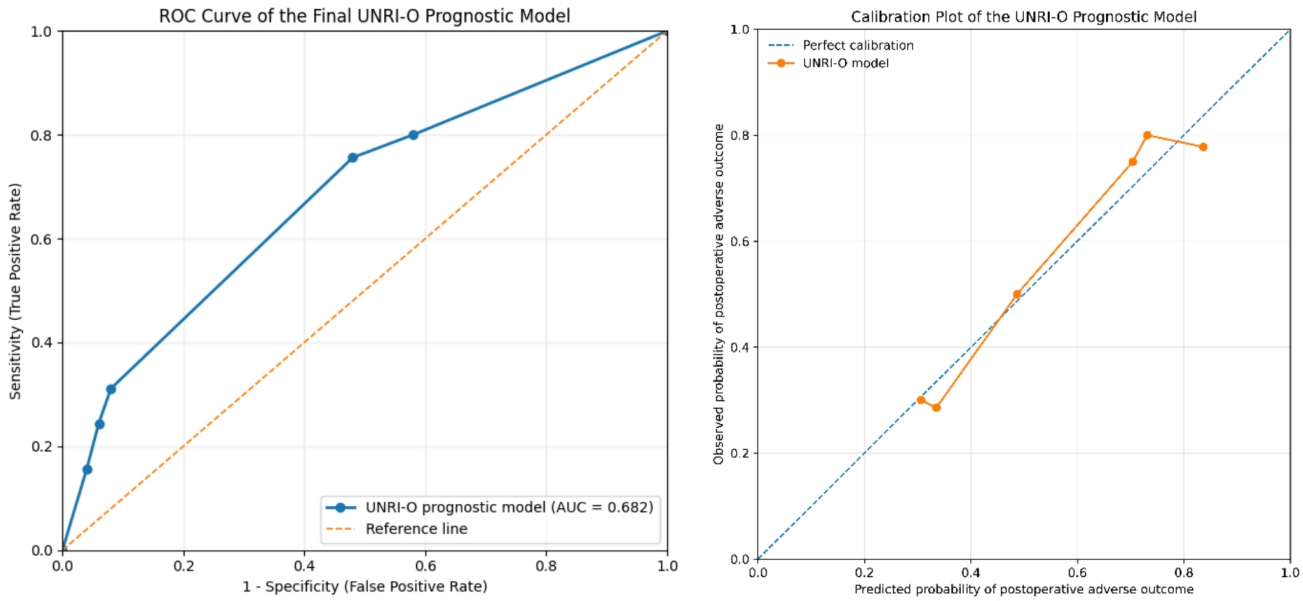


Figure 2

2A) Receiver Operating Characteristic (ROC) Curve of the Final UNRI-O Prognostic Model. 2B) Calibration plot of the UNRI-O prognostic model for postoperative adverse outcomes.

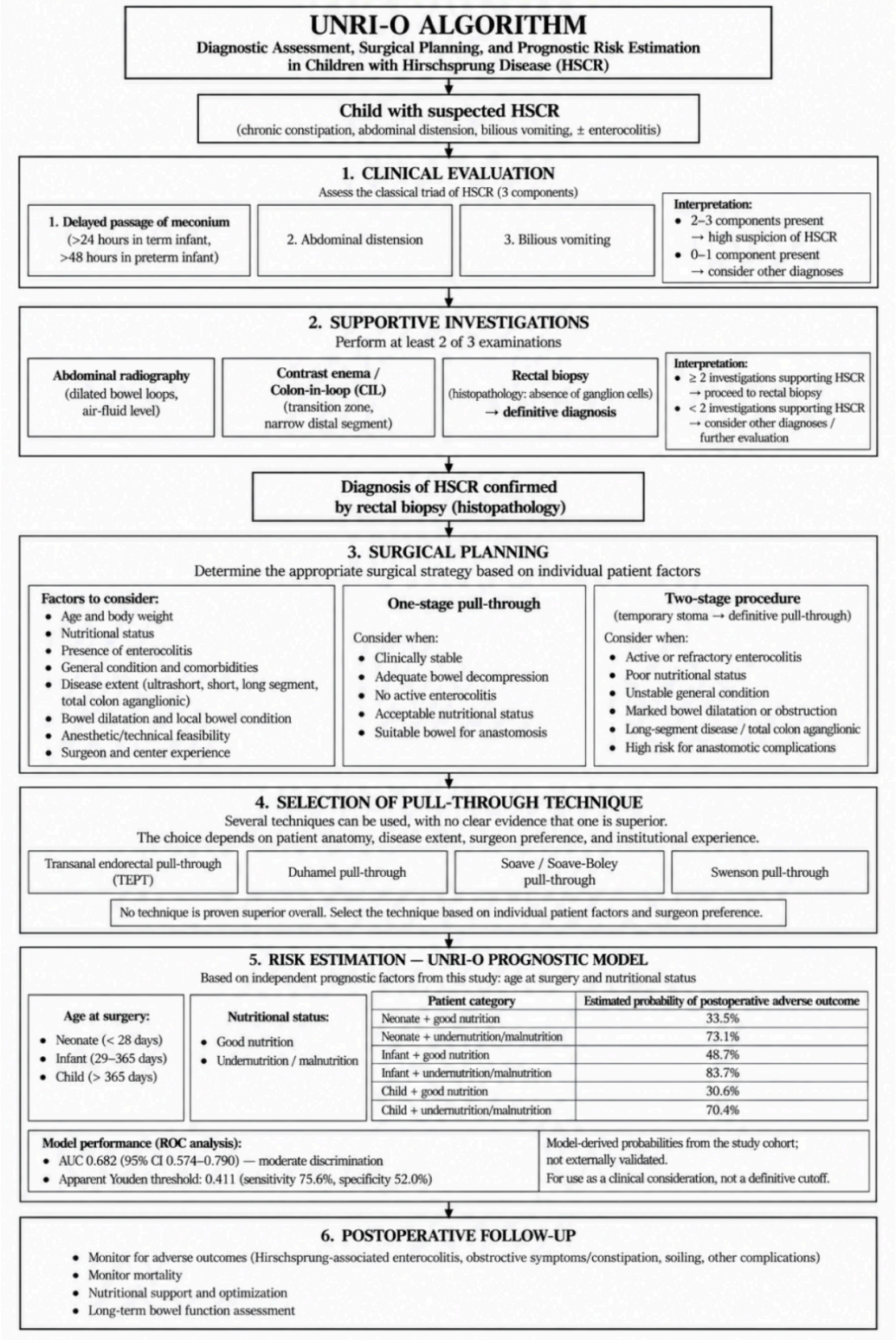


Figure 3

The UNRI-O Prognostic Risk Estimation Algorithm for Diagnostic Assessment, Surgical Planning, and Prognostic Risk.